



Case Report / Olgu Sunumu

De novo left ventricular non-compaction accompanied by severe mitral valve failure in a young pregnant

Genç bir gebede şiddetli mitral kapak yetmezliğinin eşlik ettiği de novo sol ventriküler nonkompaksiyon

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ABSTRACT

De novo left ventricular non-compaction is a rare pathology in the peripartum period. To the best of our knowledge, pregnancy-induced cardiomyopathy accompanied by severe mitral valve insufficiency requiring surgery during pregnancy has not been previously reported. Herein, we report the first postpartum case of *de novo* left ventricular non-compaction who underwent mitral valve repair.

Keywords: *de novo* left ventricular non-compaction; mitral valve repair; peripartum cardiomyopathy; pregnancy.

Peripartum cardiomyopathy (PPCM) is an unusual form of heart failure which develops during the last month of pregnancy or up to five months after giving birth. As symptoms of PPCM are nearly similar those within the normal spectrum of pregnancy and the postpartum period, its diagnosis is frequently delayed.^[1] The first series of pregnancy-induced reversible *de novo* left ventricular (LV) hypertrabeculations in primigravida females have been reported by Gati et al.^[2] The results of Gatis' study suggested that ventricular hypertrabeculations could occur in response to increased LV volume overloading or unknown physiological responses to pregnancy. Herein, we present a young lady with severe mitral valve regurgitation due to *de novo*

ÖZ

De novo sol ventriküler nonkompaksiyon, peripartum dönemde nadir bir patolojidir. Bildiğimiz kadarıyla, cerrahi gerektiren şiddetli mitral kapak yetmezliğinin eşlik ettiği gebeliğe bağlı kardiyomiyopati daha önce literatürde bildirilmemiştir. Bu yazıda, mitral kapak tamiri yapılan ilk postpartum *de novo* sol ventriküler nonkompaksiyon olgusu sunuldu.

Anahtar sözcükler: *de novo* sol ventriküler nonkompaksiyon; mitral kapak tamiri; peripartum kardiyomiyopati; gebelik.

LV non-compaction (LVNC) during pregnancy. To the best of our knowledge, we present the first case with dilated cardiomyopathy due to *de novo* LVNC accompanied by severe mitral regurgitation.

CASE REPORT

A 22-year-old postpartum patient was admitted to our hospital with worsening exertional dyspnea and palpitations 11 days after cesarean section. She had no history of cardiovascular disease and no cardiovascular risk factor such as preeclampsia or diabetes. Based on her previous echocardiographic examinations during pregnancy, she had no evidence of LVNC and had a normal ventricular function.

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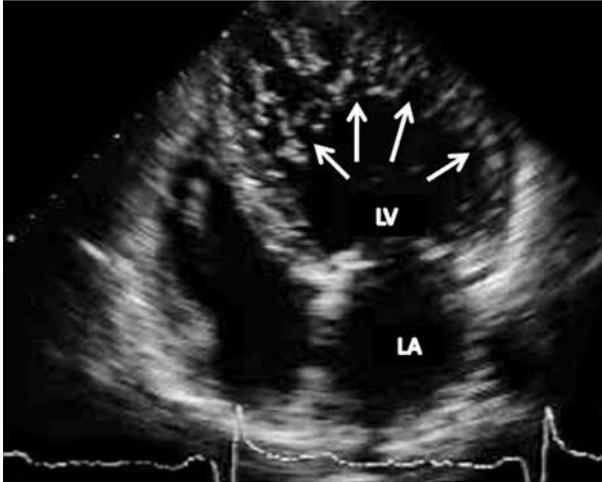


Figure 1. A view of transthoracic echocardiography showing deeply trabeculated (sponge-like) left ventricle with intertrabecular deep recesses.

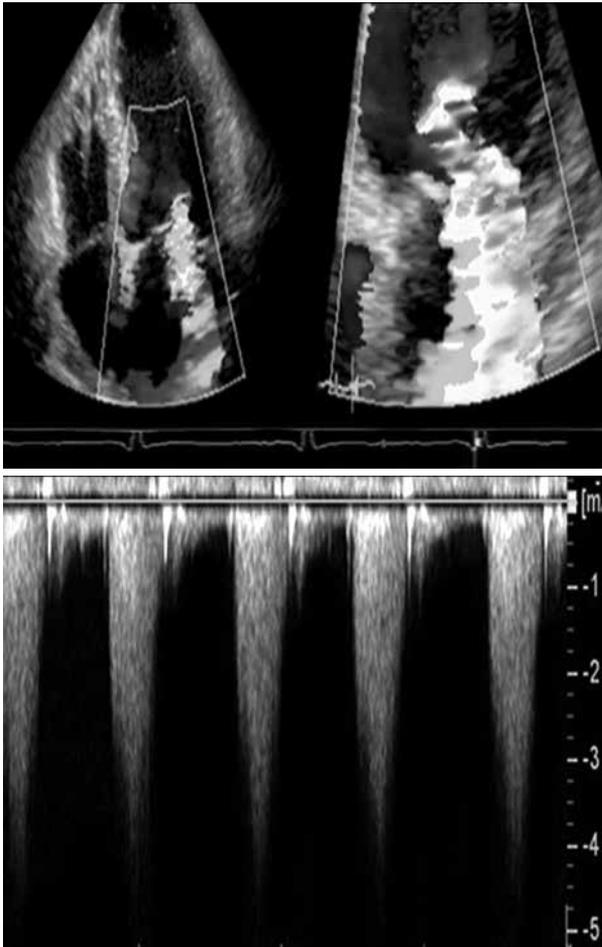


Figure 2. A view of transthoracic Doppler echocardiogram of a pregnant patient with symptomatic cardiomyopathy with severe left ventricular dysfunction (ejection fraction <25%), and Grade 4 mitral regurgitation at rest due to rheumatic heart valve disease.

In the intensive care unit (ICU), arterial pressure was 80/50 mmHg and respiratory rate was 26/min. The heart rate was 126 bpm. Cardiovascular examination revealed a Grade 4-5/6 pansystolic murmur at the cardiac apex. Transthoracic echocardiography demonstrated dilated cardiomegaly with a prominent trabecular meshwork and deep intertrabecular spaces (Figure 1). Cardiac enzymes were normal. Two-dimensional and Doppler echocardiography revealed an enlarged left atrium, and a dilated LV with a poor systolic function LV ejection fraction of 22%. There was severe mitral valve regurgitation and mild tricuspid valve regurgitation (Figure 2). The ratio of the non-compacted-to-compacted myocardial layers was 2.90 (>2.0). We performed coronary angiography and cardiac catheterization. There was a hypertrabeculated LV which constituted deep myocardial recesses (Figure 3). To relieve the symptoms, medication including levosimendan infusion was started in the ICU. Two weeks later, there was no symptom. However, no recovery of myocardial dysfunction and severe mitral valvular regurgitation were observed. Mitral valve repair was recommended to the patient.

A written informed consent was obtained from the patient. Using a midline sternotomy, aortic and venous cannulation was performed. Intraoperative findings showed severe mitral valve regurgitation and severe myxoid degeneration. We observed non-compacted endomyocardium, and LV deep recesses. Mitral valve repair was performed and a mitral valve ring was replaced. Tricuspid annuloplasty was also performed

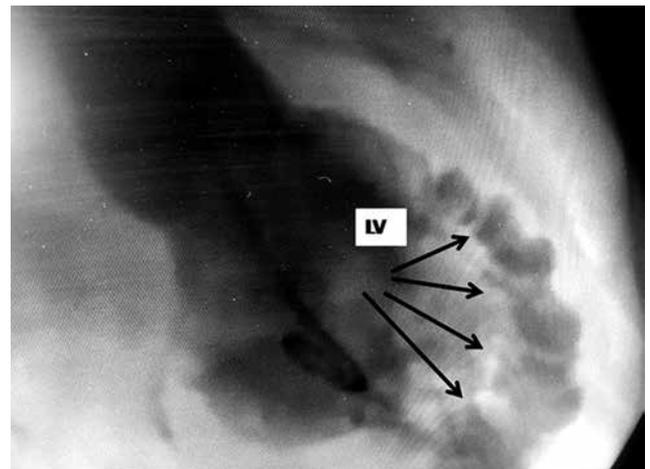


Figure 3. Left ventriculography showing a sponge-like left ventricle and deep recesses (arrows) from the view of right anterior oblique projection.

LV: Left ventricular.

using a flexible ring. The patient was discharged in a good clinical condition without any symptom. Six months later, she was still symptom-free and echocardiography showed that LV ejection fraction increased to 45% and a pulmonary artery pressure decreased to normal range.

DISCUSSION

Left ventricular non-compaction, which is a rare cause of non-classified cardiomyopathy, was first described in 1984.^[3] It is the result of an arrest of the development of myocardial compaction and is characterized by persistence of multiple, excessively prominent ventricular trabeculations, and deep intertrabecular recesses. Previously, we reported two pediatric patients with LVNC associated with congenital heart disease who underwent successful open cardiac surgery.^[3,4]

Gati et al.^[2] demonstrated that the development of *de novo* LVNC in 25% of 102 primigravida women with an age ranging between 18 and 35 years who had no evidence of LVNC or another cardiac disease in the pre-pregnancy period. In the postpartum period, they found that 18 women showed complete resolution of LV trabeculation within six months. In addition, spontaneous recovery of myocardial dysfunction in 7% of cases with LVNC was reported by Ali et al.^[6] and Gati et al.,^[2] suggested that the pregnancy induced *de novo* LV trabeculations due to LV volume overload and certain physiopathological responses. However, no patient with severe valvular heart disease has been reported previously. Therefore, to the best of our knowledge, there has been no report with cardiomyopathy accompanied by LVNC and severe mitral regurgitation after pregnancy in the literature.

The triad of LVNC is described by heart failure, arrhythmias, and embolic events which are the major clinical manifestations in patients with reduced systolic LV function. Our case was also admitted to our hospital due to LV systolic and diastolic dysfunction and sinus tachycardia.

Echocardiography is accepted as a definitive standard for the diagnosis of LVNC.^[3,5-7] Magnetic resonance imaging and left ventriculography can be also used to identify subendocardial perfusion deficits and hypertrabeculation with deep recesses.

A largest series showed that 35% of patients with LVNC died due to sudden cardiac death.^[8] A total of 12% of patients also underwent heart transplantation in the long-term follow-up period.^[9,10] Thromboembolic

events and ventricular tachycardia are the cause of poor clinical prognosis of these patients with impaired LV function. In addition, a higher LV end-diastolic diameter at the time of initial presentation, New York Heart Association Class III/IV, chronic atrial fibrillation, and bundle branch block are the most common findings of non-survivors.^[6-8]

The treatment strategies of patients with LVNC are similar to that of patients with other cardiomyopathies. Appropriate treatment includes management of arrhythmias, and oral anticoagulation to prevent systemic embolism in patients with impaired LV function.^[9,11] Early listing of symptomatic patients for heart transplantation must be seriously considered. Therefore, we performed mitral valve repair and tricuspid annuloplasty following medical management with levosimendan infusion in our case. In the literature, severe cardiovascular complications have been described.^[9]

In conclusion, we suggest that pregnancy may induce *de novo* left ventricular non-compaction. Progressive cardiomyopathy may develop in puerperium without any cardiovascular disease such as heart valve regurgitation. As *de novo* left ventricular non-compaction with symptomatic cardiomyopathy accompanied by valvular heart disease has a high mortality rate, pregnant should be followed closely and appropriate medical management should be considered. Our case with cardiomyopathy due to *de novo* left ventricular non-compaction is the first postpartum case who underwent mitral and tricuspid valve surgery in the literature.

Declaration of conflicting interests

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